Pulmonary renal syndromes

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What do the lungs and kidneys have in common?

- Detailed and extensive micro-vasculature across a large surface area
- Basement membrane (which has special antigen)
- Exchange of materials across a thin barrier.
- Both clean the body of waste product and manage the delicate balance of other materials

Facts to keep in mind

- Pulmonary renal syndromes can be **FATAL.**
- IF bleeding is occurring from both the lungs and kidneys, theses patients need to be in the ICU and get treatment **FAST** otherwise they will die
- In difficult cases with vague symptoms, sometimes an early renal biopsy can make all the difference

Definition of pulmonary - renal syndrome

- Involves combination of diffuse alveolar hemorrhage

And rapid progressive glomerulonephritis. It is usually systemic vasculitis that finally can lead to life threatening injury to lung and kidneys (Niles 1996, Salant1987,De Groot 2005)

- *Diffuse alvealar haemorrhage:* **triad of hemoptysis, diffuse alveolar infiltrate and law hematocrite**
- *Rapid progressive glomerulonephritis:* **rapid progressive renal function loss and prescence of nephritic sediments**

The basic pathology

- LUNGS:

acute onset of symptoms,2/3 cases have at least mild hemoptysis if not more gross bleeding

X-ray and CT scan are usually abnormal: alveolar and interstitial opacities, or even fibrosis

- KIDNEYS:

Acute renal failure, oliguria, RBCs cast on urine analysis

Causes of pulmonary renal syndromes

- ANCA associated vasculitis ~ 60%
  Wagners: C-ANCA
  Microscopic polyangitis:p -ANCA
  Churg- Strauss syndrome :p -ANCA
- Good pasteure's syndrome :Anti GBM ~20%
- OTHERS:

collagen disease (SLE,SS,RA)

Infections: TB, HANTA virus, legonalle, mycoplasma pneumonia, leptospirosis

Antiphospholid syndrome,TTP

-ve ANCA vasculitis :rare

- Primary renal disease can lead to pulmonary disease:

Acute renal failure with pulmonary oedema

Thromboembolis in nephrotic syndrome can lead to pulmonary embolism

Immuno -supprresion in renal disease and pneumonia

- Primary pulmonary disease can lead to renal disease:

Infection of the respiratory tract with renal affection as in IgA nephropathy

Lung cancer with immune complex nephritis

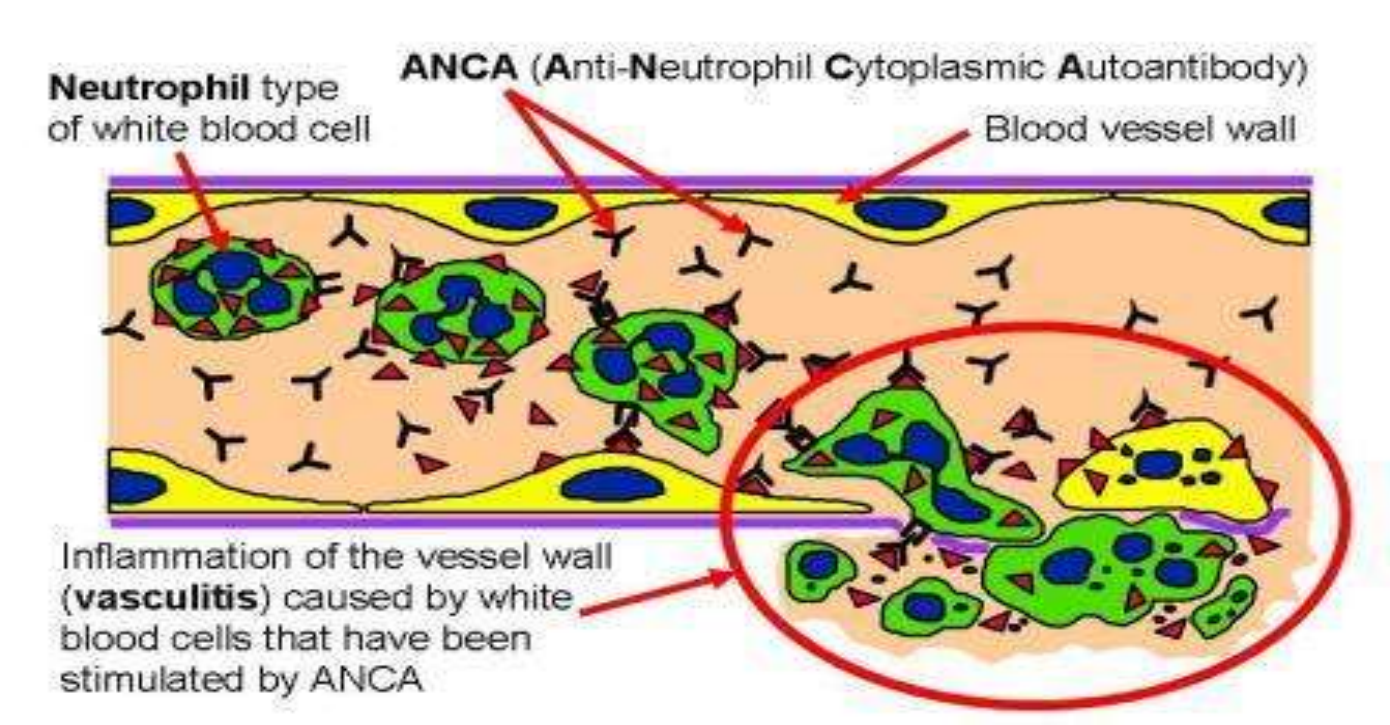
- Pulmonary renal syndrome in drug associated ANCA :positive vasculitis:

Reversible

Propyltiouracil (20% of patients) and hydralazine

D penicillamine, sulfasalazine and allopurinol

ANCA associated vasculitis



EPIDEMIOLOGY

- Begin during the 5 th, 6 th, 7 th decades of life
- Male predominance
- Caucasion have greater incidence than african americans
- Suspision of Wagners more frequent in colder compared to warmer climate and that microscopic polyangitis has opposite trend

Wagner's granulomatosis

- necrotizing vasculitis of small vessells
- Renal biopsy shows segmental necrotizing GN / rapid progressive GN in late stage
- Non caseating granuloma in lung migratory in nature
- X- ray may show: nodules, persistant infilterate,cavities
- Sign & symptoms: fever , rhinorrhea, sinus pain,sinus ulcer,hemophtysis , hematuria, proteinuria ,cutaneous purpura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A.D.A.M.


before
after

Microscopic poly angitis

- Necrotizing vasculitis, glomerulonephritis and pulmonary capiliritis
- Very similar to Wagners except no granuloma in biopsy
- Lung invoved only in 50% of the time , upper airway is rarely affected
- Used to be included with PAN –but no longer- involves the lung but PAN does not,alse is not associated with HBV, but PAN is

Churg Strauss Sundrome

- Small vessels vasculitis with focal segmental necrotizing GN
- RARE”allergic state”with systemic inflammation associated with asthma,hypergammaglobinimia, rasied IgE, and EOSINOPHIL
- SYMPTOMS :lung involvement dominates and may preceded others by years
- GN(50%),rhinitis, mononeuropathy,skin , GIT, CVS
- Renal biopsy usually does not show granuloma or eosinophils(lung and blood test show)

Treatment of ANCA vasculitis

- EUVAS group stated that patients with different levels of disease severity need different treatment protocol
- Localized, early systemic, generalized, sever, refractory
- Standered therapy by IV pulse steroid for 3-5 daya then tapring dose3-4 month to be stoped in 6-12 month
- EUVAS design the following regimen IV cyclophosphamid dose 15mg/Kg every 2 weeks for first 3 pulse then each 3 weeks for next3-6 pulse …with dose modification according to age and renal function

Treatment cont….

- Alternative for induction of remission

1- IV IG(DOSE:2 g/Kg over 5 days)

2-Infliximab (3-5 mg/kg infusion once or twice amonth)

3-MMF(2 G/DAY)

4- RITUXIMAB weekly for 4 doses

5- anti-thymocyte globulin

With treatment 85% of patient achieve remission

Goodpasture's syndrome

- Autoimmune disease with Abs against "alpha 3 non collagen domain of type 4 collagen
- 10-15% also have ANCA
- AGE: bimodal distribution
- Presentation differ acc to age
- Diagnosis:

1- renal biopsy(focal or segmetal)

2- immunoflorescence staining :antiGBM

Goodpasture syndrome

- Treatment:

Plasma pharesis
+steroid+immunosupprression

(supportive treatment if needed)

Poor prognostic criteria:

1- biopsy > 50% crescent

2- S.Cr 5-6 mg/dl

3-oliguia

4- if needs urgent dialysis at presentation

Goodpasture syndrome.....

- Even if kidney disease does not respond to plasmapharesis- lung disease does and it can be life saving
- 8-10 treatment with plasma pharesis is needed
- At same time predinisone IV PLUS cyclophosphamide should be started together
- Kidney transplant should be consider but wait about 6 month to give time for clearance of Abs

Goodpasture syndrome….

- If no treatment received 80% risk of ESRD in 1 year
- If need urgent dialysis may not ever recover renal functions
- If treatment statred early (cr<5)…one year survival will be about 90%

Goodpasture 1919

Lupus nephritis

- Immune complex deposition in glomeruli+complement cascade activation .if antiphospholipid antibodies are present then thrombotic mcroangiopathy also occur.

- In lung: pleural effusion,pleuritis ,pulmonary haemmorrhage ,uremic pulmonary syndrome

TREATMENT

- WHO classification of lupus nephritis (I to VI)

- Class I,II: no specific treatment
- Class III: mild form corticosteroid only
- Class III,IV: need induction therapy by 2-3 Gm MMF or IV cyclophosphamide
- Class V: Prednisone for 3 month and tapering dose over 1-2 year if response occur if no response discontinue the drug(if renal function deteriorate immunosuppression could be used)

Uremic lung/uremic pul. oedema

- Occurs in ESRD, especially when HTN also present
- Due to the actual uremia itself ,uremic toxins electrolyte imbalance, fluids increase pulmonary capillary permeability causing protein rich fluid to enter the lung from the capillaries causing uremic pulmonary oedema
- Xray : shows perihilar shadows with clear periphery

- Correction of uremia either by dialysis or other measure can correct the flow dynamic of pulmonary vasculature

Take home message

- Pulmonary renal syndrome is life threatening condition with acute onset and fulminant coarse if left untreated.
- Appropriate management of such patients includes accurate and early diagnosis ,exclusion of infection, close monitoring and specialized immunosuppressive treatment coupled with plasma exchange in certain cases
- New modalities of treatment could offer new hope for life saving of such patients

THANK YOU